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Scimitar Syndrome

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Scimitar Syndrome is a rare congenital anomaly characterized by an anomalous right pulmonary venous return to the inferior vena cava. It is often associated with hypoplasia of the right lung and dextroposition of the heart.¹ We report a 50-year-old man in whom the diagnosis was made incidentally during a control x-ray after a respiratory infection. The chest radiograph showed the characteristic tubular image of this anomaly (Fig. 1). The magnetic resonance image (MRI) and heart catheterization confirmed the diagnosis showing an anomalous pulmonary drainage

of the right lung into the inferior vena cava, a significant left to right shunt ($Q_p/Q_s=1:9$), and pulmonary hypertension.

Reference

1. Dupuis C, Charaf LAC, Breviere GM, Abou P, Remy-Jardin M, et al.: The "adult" form of the scimitar syndrome. *Am J Cardiol.* 1992;70:502–507.

Figure 1. Panel A: Chest radiograph showing a characteristic tubular image (arrows) coursing toward the right cardiophrenic angle corresponding to an anomalous vein drainage like a “scimitar” or Turkish sword. Panel B: Magnetic resonance angiography in a coronal projection demonstrating the presence of an anomalous pulmonary venous drainage of the whole right lung by a large tubular vein (arrow) into the inferior vena cava. Panels C and D: Heart catheterization in a selective injection into right pulmonary artery during the venous phase (Panel C) and into anomalous vein (Panel D) showing the anomalous pulmonary vein (arrows panels C and D).